CYSTIC DILATATION OF THE VERMIFORM APPENDIX.

FROM THE PATHOLOGIC LABORATORY OF THE PHILADELPHIA POLYCLINIC HOSPITAL AND COLLEGE.

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THAT cystic dilatation of the vermiform appendix is a rare condition may readily be seen when we consider that Stengel was able to collect from 9108 autopsies only 28 examples, or .3 per cent. These statistics were compiled from the following records: Steiner 2286 autopsies, 3 cases; Ribbert 400 autopsies, 6 cases; Bryant 124 autopsies, 1 case; Boody 528 autopsies, 1 case; Kelly and Hurdon 3770 autopsies, 16 cases; Stengel 2000 autopsies, 1 case. Kelynack says, " I have met with only one well-marked case of cystic appendix in an examination of several hundred cases during the past two years. No such cases are recorded in our Post-mortem Records."

After a careful review of the literature I have been able to add forty-three cases to these twenty-eight cases; and to these I wish to add one case on which this report is based. This case was observed at autopsy held in the pathologic laboratory of the Polyclinic Hospital on a case from the service of Dr. David Riesman, to whom I am indebted for the clinical notes on the case.

CASE RECORD 14006.—M. W., aged 43 years, married, colored, porter. Admitted to the medical wards April 1, 1907. Admitting diagnosis: chronic diffuse nephritis. Patient died April 7, 1907. Diagnosis: acute nephritis; uræmia. (Addendum. No history of appendicitis was given by the patient. During the physical examination there was no suspicion of the large mass afterward found in the abdomen.)

At autopsy, in addition to the kidney changes which had been appreciated before death, the following condition of the appendix vermiformis was revealed.

The cæcum is drawn over to the median line and toward the pelvis by a cystic dilatation of the appendix, which occupies the pelvis and projects upward and to the right in the ileocæcal region of the abdomen.

The specimen consisted of a cystic dilatation of the terminal portion of the appendix, having the shape of a banana, and the size of a large one, occupying the pelvis, with its proximal part toward the rectum and its distal portion pointing upward, forward and to the right into the ileocæcal region. The proximal portion of the appendix was represented by a cord about 3.5 cm. in length, the lumen of which was entirely obliterated. It rested posteriorly in the pelvis and caused marked dislocation of the head of the cæcum toward the median line. There were no adhesions connecting the appendix with any of the neighboring structures. There were no signs of an acute inflammatory process taking place. The cyst measurements are as follows: length, greatest curvature, 30 cm.; greatest longitudinal, 16 cm.; circumference, largest, 15 cm.; smallest, 13 cm.; walls, thickness, ¼ mm.; proximal obliterated portion, length, 3.5 cm.; meso-appendix extends to within 5 cm. of tip of cyst. Peritoneal surface shows marked congestion of vessels. Cyst is tense, fairly firm, and slightly fluctuant. On section there is no vestige of mucosa or submucosa. The walls are semitransparent and the inner surface is generally smooth, excepting at both ends, where there is moderate roughening and adhesions of fatty crystals and lime salts. The contents consist of a whitish-yellow, granular débris, which is in places of a yellowish, jelly-like, mucoid, gelatinous, viscid character.

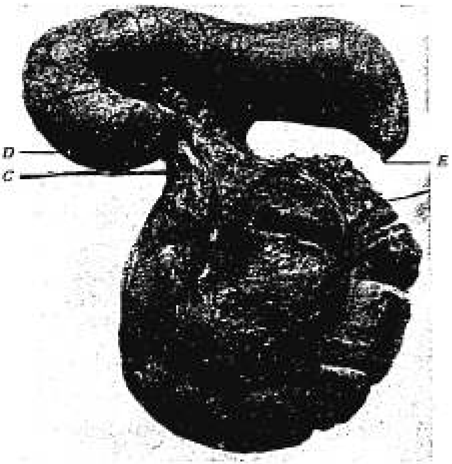
Microscopic Examination of Cyst Wall and Contents.—The cyst wall shows mucosa absent, and replaced by fibrous tissue and scattered areas of calcification. In the submucosa there are a few flattened and atrophic lymph-follicles. Serosa and muscularis are thickened and fibrous. Cyst contents show absence of definite structure, and are made up mainly of lime salts, fatty crystals, cholesterin and granular débris. No epithelial cells, leucocytes or bacteria present. Cultures taken were negative.

Historical Considerations.—Various terms have been applied to the pathologic changes which take place in cystic dilatation of the appendix. Fèré was the first to apply the term retention cyst, hydrops or mucocele to that portion of the appendix in which dilatation had occurred. The condition was first recognized by Virchow and he considered his case as one of colloid degeneration of the appendix. Since his time cases, while very infrequent, have been reported by various authorities. Most of these cases have been found during the course of operations performed on neighboring structures, and rarely for disease of the appendix itself. Many have been found during the course of a routine examination at the autopsy table. That the condition is infrequent is shown by the statements of Landenberger and others. He states: "The occurrence of cystic dilatation of the appendix is manifestly rare considering the vast number of appendectomies yearly, with the few reports of such an occurrence. Of the many surgical text-books treating on appendicitis only a few devote any space to this condition."

Etiology.—In a study of the etiology of cystic dilatation of the appendix there are a number of factors to be considered. The condition may be due to obliteration of the lumen of the appendix from internal or external lesions. Among those of internal origin may be cited: direct obliteration of the proximal end as a result of previous inflammatory changes, stenosis of Gerlach's valve, impacted fecal concretions, etc. Of the external causes there may be mentioned: angulations, adhesions causing kinking, obstruction due to bands, etc.

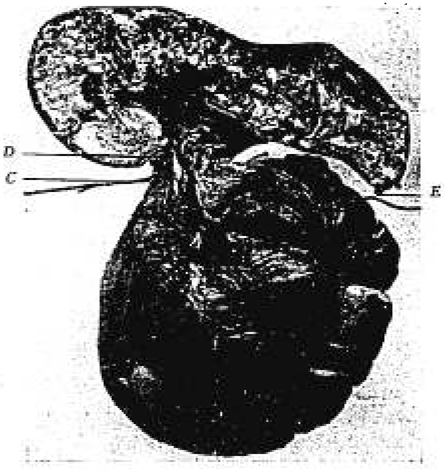
Ribbert in formulating the changes which take place in an appendix where cystic dilatation occurs, states as follows: "In the lesser degrees of dilatation we see no essential alteration in the composition of the wall. Later, abnormalities of the epithelium occur; it is lost, together with the glands, throughout considerable areas, and forms in other parts a covering for a single layer without glands, which still, however, may be present in patches; in very great dilatation it may be destroyed altogether, but disappears when dilatation is slight. We must, however, take into consideration that the loss of the epithelium does not need to be the result of dilatation, but that it may be destroyed by the disease process, which led to

FIG. 1.



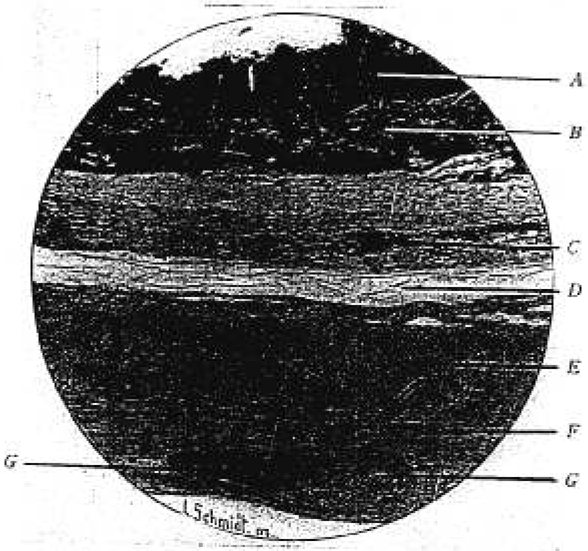
Cystic dilatation of the appendix vermiformis. Outer view. *A*, distal dilated portion of the appendix; *B*, cæcum; *C*, proximal portion of appendix; *D*, point of occlusion; *E*, tip of appendix.

FIG. 2.



Cystic dilatation of the appendix vermiformis. Inner view. *A*, distal dilated portion of the appendix; *B*, cæcum; *C*, proximal portion of appendix; *D*, point of occlusion; *E*, tip of appendix.

Fig. 3



A transverse section through the wall at the seat of the dilatation. *A*, lime-salts on surface of mucosa; *B*, remains of mucosa: *C*. atrophic lymph follicles; *D*, œdema of mucosa; *E*. circular, and *F*. longitudinal muscular fibres showing fibrosis; *G*, greatly distended artery and vein in serosa. × 70.

the cicatrization producing the obliteration of the lumen. The follicles disappear equally soon, and finally are altogether wanting."

Bischoff thinks that a dilatation is wanting when the mucous membrane is still able to resorb in the normal way. Ribbert maintains that a dilatation is also wanting when as a result of early and extensive destruction of the mucous membrane, no secretion in the lumen occurs.

Latham states that "a number of factors may lead to retention of its secretion in an inflamed appendix. When its muscular coat is inflamed, and its functional activity thus impaired, the expulsive powers of the appendix become insufficient. In other cases external mechanical conditions such as twists, kinks, abnormal position or fixation of the organ give rise to retention. The swelling of the mucosa itself and Gerlach's valve may also prevent the exit of the contents of the appendix. The secretion after it has once begun to accumulate favors further retention in two ways, *i.e.*, in the first place by exerting mechanical pressure on the wall of the appendix; in the second place by serving as a good culture medium for the growth of bacteria."

Obermayer states: "In a minority of the cases the process terminates in a different condition which is known as hydrops of the vermiform appendix (mucocele). In these cases the lumen is locally obliterated, presumably as a result of ulcers due to catarrhal ulcerative appendicitis (with or without stercoliths), or occasionally by kinking or constriction of the appendix by external factors, chiefly chronic peritoneal adhesions. These internal cicatrices are comparatively common near the cæcal orifice, but they may be found in any part of the organ; this also applies to constrictions due to external causes. The cicatricial stenosis is not always complete, and there is sometimes a minute communication between the lumen of the appendix and the cæcum when obliteration of the lumen occurs in the appendix; when no virulent bacteria are present, no suppuration occurs. Cystic dilatation of the distal part of the organ does not necessarily follow, and does not occur when the secretion present in the organ is absorbed or when the mucous membrane is destroyed, and therefore, unable to manufacture any further secretion."

Deaver states that, " Not all cases are due to an obliteration of a portion of the lumen of the appendix. Sufficient obstruction may be produced by an acute angulation. The development of this condition of cystic dilatation of the appendix depends upon several factors. The obstruction of the lumen must be complete, or almost so; the obstruction or obliteration must have obtained at a time when the affected portion of the organ contained no pathogenic micro-organisms; the mucous membrane of the affected portion must be intact, or at least capable of functionating, and the secretion by the mucous membrane must be more rapid than the absorption from the portion of the appendix involved. If the obstruction be not complete, the secretion is likely to be forced through even a narrow opening with sufficient rapidity to prevent a large accumulation, and on the other hand, infection is likely to occur through the patulous lumen, converting a cystic dilatation into an empyema. If there are already virulent bacteria in the affected portion of the appendix, an empyema, of course, rather than a cystic dilatation, will develop in the first place. If the mucous membrane is incapable of functionating there can be no accumulation of fluid, and the same is also true if absorption be more rapid than secretion."

Kelly and Hurdon state, " If a portion of the canal distal to the obstruction remain patent, the normal secretion, having no outlet, accumulates in it and a retention cyst is produced. The contents of the cyst are, at first, composed of the normal mucous secretion of the appendical mucosa, or of a mucopurulent, sometimes sanguinous fluid; but later, probably owing to pressure atrophy of the mucosa and consequent loss of function, the mucous secretion ceases, and the fluid becomes serous or watery in character. If the occlusion has occurred close to the cæcal attachment of the appendix, the cyst appears to arise directly from the cæcum. If the obstruction is at a more distant point the cyst appears to be pedunculated, being attached to the cæcum by the normal proximal portion of the appendix."

Gross Pathology.—Cysts of the appendix vary greatly in size. Cases have been reported which have ranged in size from a pea to a large walnut, while exceptional ones have been as large as a pear or even greater,—such as that described by Sonnenberg, which measured 14 cm. in length and 21 cm. in its greatest circumference. Other large cysts have been reported by Virchow and others. Virchow's specimen was the size of a fist. Deaver reports a specimen the size of a small orange. The shape of these cysts also varies. They may be cylindrical, ovoid, or irregular. As a rule where the occlusion is at the junction of the cæcum and appendix, they are symmetrically dilated, usually being sausage- or banana-shaped. When the occlusion is situated some distance from the cæcum, they may be small and round. Occasionally secondary cysts develop in the remains of partly occluded glands. Hernia-like protrusions into the mesentery may occur, as in the cases described by Ribbert, Kelynack, and Kelly and Hurdon. At times the cyst is produced by bands of adhesions at the cæcal end, and when this takes place, and although the cyst is very large before operation, its size may suddenly diminish during exposure on account of the manipulations necessary to bring it into view, thus causing the adhesions to break or the kink to be released. As a result the cyst contents escape into the cæcum, and its walls become flaccid.

The external appearances of the cysts are more or less uniform. As a rule there are few if any adhesions connecting the cyst with surrounding structures. The walls are smooth, firm, thickened and transparent. Dilated enlarged blood-vessels can be seen in their walls. The inner surface is generally smooth, glistening, and attenuated. At times there is a well-marked deposit of lime salts upon its inner surface, and its contents, if organized, may in places be firmly attached to its walls. At times the contents may be clear, watery, serous, and of a whitish or yellow color. When this condition is present the term hydrops of the appendix may be applied.

Generally, however, the contents are of a whitish-yellow, slightly turbid, gelatinous character. The contents may at first be of a tenacious mucoid character and later change to a serous nature. Deaver states that "this ensues because the wall of the appendix, as it becomes distended, usually also becomes thinned, its inner surface growing smooth and the distribution of its vessels being more superficial than normal. This results on the one hand in facilitating the escape of the watery parts of the blood, and on the other, in reducing to a minimum the formation of the mucus."

Microscopic Pathology.—The microscopic appearances of the wall of the cyst generally consist of moderate chronic inflammatory changes associated with pressure atrophy of its walls. The mucosa is generally thinned and may be entirely absent, or in a moderate degree of atrophy, due to the mechanical pressure of the contained fluid. The submucosa may be œdematous. The lymph-follicles are either very much flattened or entirely absent, dependent, of course, upon the size of the cyst and the length of time the process has been progressing. Generally the muscular layers are well preserved, but there is usually a moderate amount of fibrous tissue present. At times the muscular layers are entirely replaced by fibrous tissue. When the process has been moderately slow and there has been a mild grade of inflammation present, the walls may be more thickened than normal. The contents of the cyst are generally sterile.

Résumé of the Cases from the Literature.—A summary of the cases found in the literature and that of the author, in all 68 cases, shows the following facts: 33 cases were observed at the operating table, 18 were found at autopsy, and in 17 the method of observation is not recorded. Of the 33 cases observed at the operating table there were symptoms of appendicitis in 20. In 13 cases the cyst of the appendix was found during the course of operations upon other structures. Of these 13 cases, 4 were present in hernial sacs, and 2 were associated with ovarian cysts. Of the 68 cases, 9 were found in males, 8 in females, and in 51 the sex is not recorded. The cyst included the entire appendix in 11 cases, it was localized to a portion of the organ in 20, and in 37 no note was made as to whether it was general or local. The contents were described as being mucoid in 4 cases, gelatinous in 12, colloid in 4, fluid in 7, and in 4 cases there were carcinomatous areas present in the walls of the cysts. Of the 32 cases in which the age is recorded it is given as follows: between 1 and 10 years, one case; between 11 and 20 years, two cases; between 21 and 30 years, seven cases; between 31 and 40 years, eight cases; between 41 and 50 years, seven cases; and between 51 and 60 years, seven cases. While the great majority recorded were not very large, some were notably so, such as the following cases: Wood's and Combemale's were 20 cm. in length; Finkelstein's measured 16 cm. by 14 cm.; Montgomery's measured 5½ in. by 4¾ in.; the author's measured 30 cm. along its greatest longitudinal curvature and 14 cm. in its greatest circumference; Deaver's was the size of an orange; and the one recorded by Virchow was the size of the fist.

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